The microscopic picture of ILVEN is essentially that of a non-specific chronic dermatitis(6). It is characterized by moderately well circumscribed, short and broad columns of parakeratosis, arising from epidermis and are perpendicular to the skin surface usually at the summits of epidermal papillation. Between the areas of parakeratosis the stratum corneum is hyperkeratotic in compact patterns. Beneath the zones of parakeratosis, granular layer and dyskeratotic or vacuolated are absent and beneath the areas of orthokeratosis, granular layer is thickened. The epidermis is hyperplastic, and usually papillated and psoriasiform. Superficial perivascular lympho-histiocytic infiltrate is seen in the dermis(1,3,4,7-9). In a few lesions, extensive parakeratosis is seen(2,5).

Older way of treatment in the form of topical corticosteroids, surgical excision or cryosurgery was not satisfactory. Considerable improvement in the lesion and delayed recurrence of inflammatory episodes were observed by local application of potent corticosteroids like betamethasone dipropionate(3).

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CYSTIC KIDNEYS IN TUBEROUS SCLEROSIS

S.R. Ranade
P.A. Amanapure
D.V. Raichur
P.G. Patil

Tuberous sclerosis is a neurocutaneous disorder which in addition to the skin and brain affects the kidneys, heart, eyes, lungs and bones. Nearly 50 to 80% of the patients with tuberous sclerosis have renal angio-

From the Department of Pediatrics, Wanless Hospital, Miraj.

Reprint requests: Dr. (Mrs) Shashikala R. Ranade, Head of Department of Pediatrics, Wanless Hospital, Miraj 416 410.

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myolipomata. Renal cysts are recognized to be unusual manifestation of the disease(1).

So far we have not come across any case reports in Indian literature describing multiple cystic renal lesions in tuberous sclerosis, although Dewan and Agarwal described a case of tuberous sclerosis having multiple hamartomas and a well circumscribed, single, large, cystic angiolipoma in one kidney(2). We report a case of tuberous sclerosis with bilateral multiple renal cysts, which so far has not been reported in the Indian literature.

Case Report

An 18-month-old boy was brought with history of convulsions since 9 months. The convulsions were generalized, clonic, associated with deviation of eyes, lasting for 5 to 6 minutes and occurring 4 to 5 times in a day. The patient had history of delayed motor and language development. There was no history of birth asphyxia or head injury. He was being treated with phenobarbitone by a local practitioner who referred the patient to us after finding a mass on right side of the abdomen. The family history was non-contributory.

On examination, the patient had anthropometric values within normal limits, was afebrile and had a pulse rate 110/min and blood pressure 100/60 mm Hg. He had adenoma sebaceum over both malar areas, measuring 1 to 4 mm in size. Multiple hypopigmented macules and patches of size 0.5 to 3.0 cm were present on anterior and posterior aspects of the trunk. An elevated flesh colored plaque, shagreen patch, was present on upper part of the abdomen. Ophthalmoscopic examination was within normal limits. There were no focal neurological deficits. Developmental milestones were delayed. On palpation of abdomen there were lobulated, firm flank masses of size 10 cm × 7 cm on right side and 12 cm × 5 cm on left side. Liver and spleen were not enlarged. Respiratory and cardiovascular systems were within normal limits.

On investigation, the levels of blood urea, creatinine and hematological profile were within normal limits. Urine examination did not reveal any abnormality. Roentgenograms of chest and skull were normal. Abdominal ultrasonography (*Fig. 1*) revealed enlarged kidneys bilaterally with multiple cysts. Intravenous pyelogram showed enlarged kidneys with intermittent extrinsic compression and stretching of both pelvicalyceal systems, suggesting "adult type polycystic kidneys".

A diagnosis of tuberous sclerosis with multiple renal cysts was made. The child was treated with sodium valproate and nitrazepam, for control of convulsions.

Discussion

Tuberous sclerosis is inherited as an autosomal dominant trait and has wide variation in expression. Of all cases, 50 to 70% may arise from new mutations(3). Criteria for the definite diagnosis of tuberous sclerosis include one or more of the following conditions: facial angiofibroma, ungual fibroma, retinal hamartoma, cortical tuber, subependymal glial nodule and renal angiomyolipomata. Criteria for a presumptive diagnosis of tuberous sclerosis include two or more of following conditions: hypomelanotic macule, shagreen patch, cardiac rhabdomyoma, gingival fibroma, pulmonary lymphangiomyomatosis, dental enamel pits, peripapillary retinal hamartoma, infantile spasms, a single renal angiomyolipoma, multicystic kidney, "honey comb" lung on radiographic examination, myotonic, tonic or atonic seizures and an immediate relative with tuberous sclerosis(4).

Cystic kidneys are associated with various malformation syndromes. However,

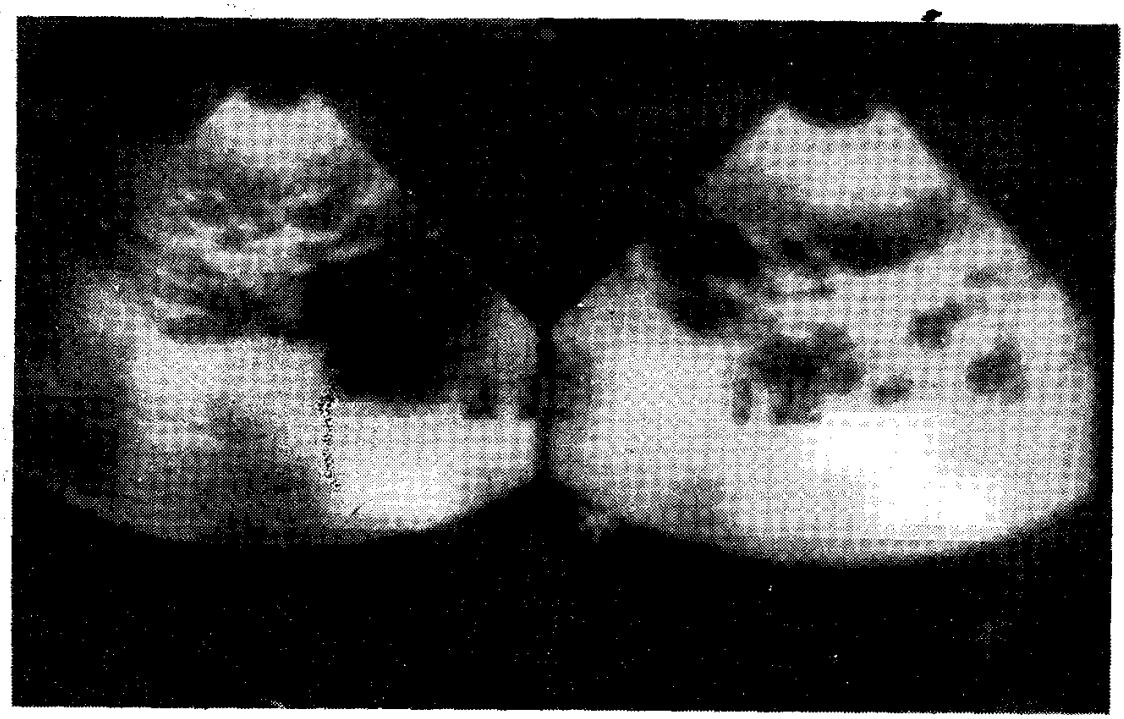

Fig. 1. Ultrasonogram demonstrating bilateral multiple renal cysts.

external manifestations present in these cases distinguish them from adult type polycystic kidneys(1,5). In our case though abdominal ultrasonography and intravenous pyelography suggested adult type polycystic kidneys, the presence of delayed developmental milestones, seizures and characteristic skin lesions pointed to the diagnosis of tuberous sclerosis with multiple renal cysts. Even in the absence of other manifestations, mental retardation and seizures when associated with cystic kidneys suggest a diagnosis of tuberous sclerosis(1). However, in some cases, cystic renal lesions may be the presenting feature of tuberous sclerosis, even before other manifestations develop. Therefore, large cystic kidneys in infancy with normal hepatic anatomy and function and without family history of polycystic kidney disease should lead us to the possibility of tuberous sclerosis(6). In such cases a renal biopsy is indicated(1). Polycystic kidneys when occur in early childhood usually are associated with a bad prognosis due to renal failure. In case of multiple renal cysts of tuberous sclerosis, early renal failure does not occur(7). However, hypertension, hematuria, pain abdomen and rarely renal failure do manifest(1,8,9).

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PRECOCIOUS PUBERTY, GELASTIC SEIZURES AND HYPOTHALAMIC HAMARTOMA

P.S.N. Menon
N.K. Mishra
V. Muthukumar

Hypothalamic hamartoma is a well defined rare syndrome which often presents with precocious puberty and seizures(1). With the advent of magnetic resonance imaging (MRI), even very small sized hamartomas can be picked up. Using gonadotropin releasing hormone (GnRH) analogs, precocious puberty as well as skeletal maturation can be controlled without any major side effects(2). However, the seizures which occur because of cerebral dysgenesis still pose a therapeutic problem(3). We report here a boy with hypothalamic hamartoma who presented with precocious puberty and gelastic seizures.

Case Report

A product of full term normal delivery following an uneventful antenatal period presented at 2½ years of age in 1985 with increasing size of testes and penis noticed immediately after birth and increased appetite from early infancy. Pubic hair appeared at 6 months of age and erections and ejaculations were observed at 1 yr of age. There was no axillary hair. In addition, he had behavioral abnormalities in the form of obstinacy and temper tantrums.

He was investigated in our hospital in 1985. CT scan and CT cisternography revealed a suprasellar isodense mass suggestive of hypothalamic hamartoma (*Fig. 1*). He underwent surgery in 1986 with a right frontal craniotomy with partial decompression of sella. Follow up CT scans showed persistence of the tumor. He was put on buserelin, a GnRH analog for arresting further pubertal development.

Six months later he developed secondary tonic seizures. Acne appeared at the age of 4½ yrs and chest hair appeared at 6½ yrs. He had discontinued buserelin therapy after one year due to financial problems. In 1991, at the age of 8½ yrs, he again

From the Division of Pediatric Endocrinology, Department of Pediatrics and Department of Neuroradiology, Neurosciences Centre, All India Institute of Medical Sciences, New Delhi 110 029.

Reprint requests: Dr. P.S.N. Menon, Additional Professor of Pediatrics, All India Institute of Medical Sciences, New Delhi 110 029.

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From the Division of Pediatric Endocrinology, Department of Pediatrics and Department of Neuroradiology, Neurosciences Centre, All India Institute of Medical Sciences, New Delhi 110 029.

Reprint requests: Dr. P.S.N. Menon, Additional Professor of Pediatrics, All India Institute of Medical Sciences, New Delhi 110 029.

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